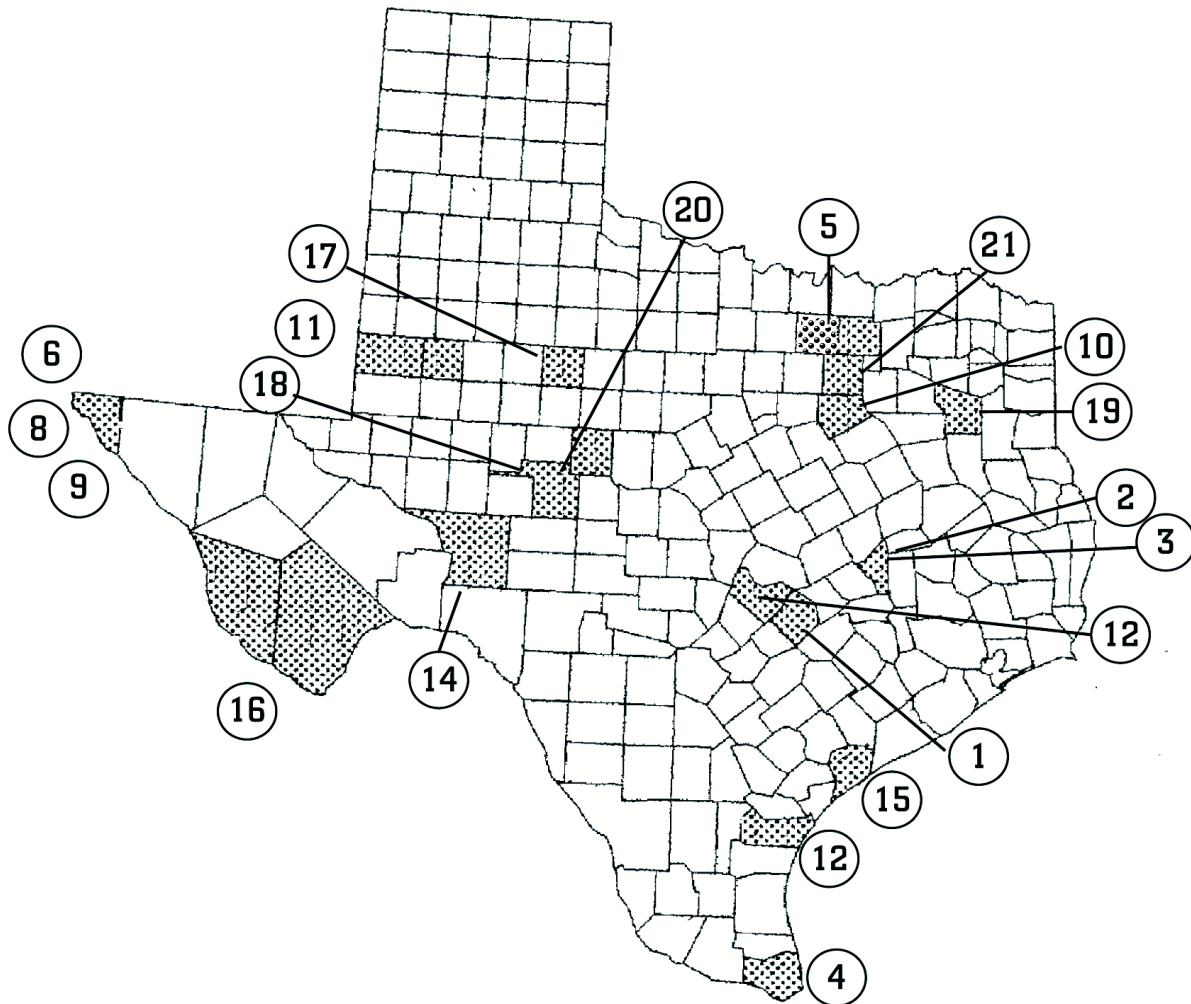


Birth Defect Investigations Conducted in 1999

By the Texas Birth Defects Monitoring Division,

Texas Department of Health



See report below for corresponding numbers.

Introduction

Health care professionals, parents, and others can report apparently unusual concentrations of birth defects to the Texas Birth Defect Monitoring Division (TBDMD). Birth defect cluster investigations are then initiated. A glossary defining the birth defects mentioned in this report is available by phoning 512-458-7232.

Methods

Cluster investigations follow a protocol that helps us spend more time on those investigations that are likely to provide useful results.¹ For example, say someone reported to us that there were fifteen children born with cleft lip/palate in 1998 to mothers who resided in Travis County. Since we know that birth defects occur everywhere at a certain background level, the question we ask is not "Should the cases have occurred at all?", but more "Were there more cases seen than we would have expected?". To answer that question, we consider two things - the background level, and the role of chance.

We would use the whole State of Texas to determine the background level of birth defect occurrence, but do not yet have those data. So we usually use the rates experienced in the oldest active birth defects registry in the US, the Metropolitan Atlanta Congenital Defects Project (MACDP) or the largest registry in the US, the California Birth Defects Monitoring Program (CBDMP). We then ask for example, "If the 12,614 births in Travis County in 1998 had the same frequency of cleft lip/palate as they see in California, how many cleft lip/palate cases would we expect in Travis County that year?" The answer is 13.88 cases.

But we know that even if we expect 13.88 cases per year, the number that will be born will bounce around; in some years it will be 13 cases, in some 10 cases, and in others 17 cases, etc. This is where the role of chance comes in. We can do calculations to tell us the chance of getting 15 cases when we expect 13.88. (This is similar in principle to calculating the chance in coin flipping of getting 15 heads in a row when we would expect 7.5 heads.) It turns out that by chance, 15 cases of cleft lip/palate would turn up about 4 out of 10 times. That's pretty common, so we conclude that chance could easily explain this occurrence. If the chance is less than 1 out of 20 times (less than 0.05), we feel that's really stretching it, so we customarily say that chance is probably not the only explanation for what's going on here; another term for that is "statistically significant".

"Ascertain" means to find and record. We can ascertain cases of birth defects in many ways, such as surveying parents or service providers, looking through birth certificates, or systematic searching through records in hospitals.

Cluster Investigations in 1999

In the calendar year 1999, the TBDMD investigated a total of 21 potential birth defect clusters. Of 12 initiated in previous years, 4 were concluded in 1999 (items 2, 3, 4, 11). Of 9 initiated in 1999, 1 was concluded (item 5). Altogether, 16 investigations will continue into 2000.

Nearly one quarter of the reports investigated concerned neural tube defects (items 1, 3, 4, 5, and 8). Approximately 14% of the reports concerned oral clefts (items 11, 14, 16). Heart

¹ For a copy of the protocol, contact Peter Langlois at (512) 458-7232.

defects (items 2, 18) and chromosomal defects (items 9, 19) each accounted for 10% of the investigations.

Most of the reports of suspected birth defect clusters investigated during 1999 were reported by health care or service providers.

(1) BASTROP COUNTY

Condition of Concern: *Anencephaly*

Time Period of Concern: Unknown

Background: A parent called with a concern about excessive anencephaly in the area.

Response: TBDMD staff are working with the parent to determine the number of cases reported, time period, and geographic area of concern. This investigation is ongoing.

Duration of Investigation: October 1999 - December 1999

(2) BRAZOS COUNTY

Conditions of Concern: *Hypoplastic left heart syndrome*

Time Period of Concern: July 1997 - December 1997

Background: In May of 1998, TBDMD was contacted by a citizen of Brazos County about a perceived high number of infant deaths due to hypoplastic left heart syndrome in the county. Three cases were reported.

Response: TBDMD confirmed a total of three cases of hypoplastic left heart syndrome in Brazos County in the time period of interest. Based on CBDMP rates, this was 11.9 times the expected number of cases and was statistically significant. However, since only three cases were involved, further investigation to try to find the cause of this excess occurrence is unlikely to be fruitful. Consequently, no further action is required at this time.

Duration of Investigation: May 1998 - April 1999

(3) BRYAN AND COLLEGE STATION (Brazos County)

Condition of Concern: *Anencephaly*

Time Period of Concern: 1981 - 1996

Background: A resident of College Station contacted the Texas Department of Health

with concerns about anencephaly. He stated that there had been 20 pregnancies affected by anencephaly in his neighborhood. No specific time period of concern was mentioned.

Response: Vital records (birth, fetal death, and death certificates) from 1981 to 1996 for residents of Brazos County were searched to identify all pregnancies affected by anencephaly. Six affected pregnancies were identified. Five of those were born to residents of Bryan and one to a resident of rural Brazos County. No affected pregnancies were detected among residents of College Station. There was no statistically significant excess of anencephaly observed in Brazos County, the city of Bryan or the city of College Station from 1981 through 1996.

After TBDMD reported these findings, the investigation was reopened when a concerned parent provided the names of 5 families believed to have had a pregnancy affected by anencephaly. Birth, death, or fetal death certificates were found for 4 of these 5 children. Two children had been identified in the initial investigation. One child was not included in the initial investigation because the child's cause of death was listed as encephalocele, rather than anencephaly. One child was not included because the mother was listed as a resident Travis County, rather than Brazos County. No further action is required at this time.

Duration of Investigation: March 1997 - October 1999

(4) CAMERON COUNTY

Condition of Concern: *Neural tube defects*

Time Period of Concern: February 1998 - December 1998

Background: A staff member from a local newspaper contacted the Texas Department

of Health about a possible increase in infants born affected by neural tube defects (NTDs) in Brownsville and elsewhere in Cameron County in 1998. He reported that 9 affected pregnancies had been identified from February through July.

Response: Case finding and verification were undertaken using data from the Texas Neural Tube Defect Surveillance and Intervention Project (TNTDP). A total of 17 affected infants or fetuses were confirmed as having been delivered from February through October 1998. The excess occurrence in Cameron County was statistically significant and it ended a downward trend observed there since 1994. An investigation into what might cause high NTD rates in all Texas counties bordering Mexico is ongoing with the TNTDP. The TNTDP also works to prevent a second NTD-affected pregnancy among their study subjects. Texas Department of Health staff are working with partners to develop and implement a campaign to prevent the first occurrence of an NTD in families. No further action on this cluster investigation is required at this time.

Duration of investigation: July 1998 - May 1999

(5) COLLIN AND DENTON COUNTIES

Conditions of Concern: *Neural tube defects*

Time Period of Concern: January 1997 - November 1997

Background: In November of 1999, staff of the Texas Birth Defects Monitoring Division in the Dallas/Fort Worth region reported what they perceived to be a higher than expected number of neural tube defect cases born to residents of Collin and Denton Counties.

Response: Using data from the Texas Birth Defects Registry, a total of 7 confirmed cases of neural tube defects were identified as being delivered to residents in the two counties during the time period of interest. Based on CBDMP rates, this was 0.7 times the expected number of cases and not statistically significant. Since there was not a higher than expected number of cases, no further action is required at this time.

Duration of Investigation: November 1999

(6) EL PASO (El Paso County)

Condition of Concern: *Biliary atresia*

Time Period of Concern: 1992 - 1997

Background: A parent reported that 6 babies with biliary atresia had been born in El Paso from 1992 - 1997. He was concerned that this was excessive. Based on rates from CBDMP, this was within the limits of what would have been expected. However, the parent felt that there may have been more affected children than those he had reported.

Response: Further case finding and case verification will be conducted using data from the Texas Birth Defects Registry for deliveries in 1997. This investigation is ongoing.

Duration of Investigation: September 1997 - December 1999

(7) EL PASO (El Paso County)

Condition of Concern: *Multiple defects*

Time Period of Concern: 1994 - 1997

Background: A physician contacted TBDMD, concerned that he was seeing an abnormally high number of children with hypoplastic left heart syndrome, various intestinal interruptions, and cancer.

Response: Further information is being gathered.

Duration of Investigation: May 1997 - December 1999

(8) EL PASO (El Paso County)

Condition of Concern: *Neural tube defects*

Time Period of Concern: September 1999 - October 1999

Background: Regional staff of the Texas Birth Defects Monitoring Division reported concern that 7 pregnancies with a neural tube birth defect were delivered during a two month period to mothers living in the city of El Paso. Five pregnancies were affected by anencephaly. This was approximately 4 times the number of anencephaly cases that would be expected and was statistically significant. Spina bifida (2 cases reported) and total

neural tube defects (7 cases reported) were not statistically significantly elevated.

Response: Since the geographic area and time period of concern are covered by the Texas Neural Tube Defect Project (TNTDP), data from the TNTDP will be used to confirm reports and ascertain any additional cases among residents of El Paso. The investigation is ongoing.

Duration of Investigation: November 1999 - December 1999

(9) EL PASO (El Paso County)

Condition of Concern: *Trisomy 18*

Time Period of Concern: January 1999

Background: A hospital nurse reported her concern about 2 children with Trisomy 18 born in a single month. The mothers of both children lived in the city of El Paso. Based on rates from CBDMP, this was 9 times what would be expected and was statistically significant.

Response: Since the geographic area and time period of concern are covered by the Texas Birth Defects Registry, registry data will be used to confirm reports and identify any additional cases among El Paso residents. Collection of registry data for 1999 is still underway, therefore the investigation is ongoing.

Duration of Investigation: February 1999 - December 1999

(10) ELLIS COUNTY

Condition of Concern: *Multiple defects*

Time Period of Concern: March 1993 - August 1998

Background: Staff at Baylor Medical Center in Ellis County were concerned with what may have been high numbers of birth defects among deliveries at their hospitals. They contacted the Texas Birth Defects Monitoring Division to discuss their concerns.

Response: We reviewed the report that was provided and have requested further information on how their analyses were performed. The investigation is ongoing.

Duration of Investigation: October 1998 - December 1999

(11) GAINES AND DAWSON COUNTIES

Conditions of Concern: *Cleft lip and cleft palate*

Time Period of Concern: 1991 - 1996

Background: In March of 1995, a parent contacted TBDMD about a potential excess of cleft lip and cleft palate. Three affected infants were reported as having been born to Gaines County residents in 1994-1995. Based on CBDMP rates, this was 16.0 times the number expected and was statistically significant. The investigation was expanded to include neighboring Dawson County when excessive numbers of babies born with cleft lip and/or palate were reported there.

Response: TBDMD identified a total of 11 infants with cleft lip and cleft palate delivered to residents of either Gaines or Dawson Counties from 1991-1996. Based on CBDMP rates, this was more than twice the number expected and was statistically significant, both alone and when adjusted for race/ethnicity. Most of the excess appeared to be due to excessive cleft lip with or without cleft palate in Gaines County. A feasibility study was reviewed by TBDMD epidemiologists and selected members from the Scientific Advisory Committee for Birth Defects in Texas. Based on that, it was decided not to do a case-control study because there were too few cases, exposure was too long ago, interview studies often do not address environmental concerns adequately, and it would be too costly. TBDMD will keep a close eye on oral cleft occurrence in these counties. No further action on this cluster investigation is required at this time.

Duration of Investigation: March 1995 - November 1999

(12) NORTH AUSTIN (Travis County)

Conditions of Concern: *Gastroschisis and omphalocele*

Time Period of Concern: Unknown

Background: In November of 1999, an obstetrician reported apparently higher than expected number of gastroschisis and/or omphalocele cases in North Austin.

Response: TBDMD is waiting for the physician to provide the number of cases and time period in an effort to perform initial calculations. The investigation is ongoing.

Duration of Investigation: November 1999 - December 1999

(13) NUECES COUNTY

Conditions of Concern: *Anencephaly and cyclopia (holoprosencephaly).*

Time Period of Concern: February 1998 - October 1998

Background: In May of 1999, regional TBDMD staff reported what appeared to be a higher than expected number of cases of anencephaly or cyclopia (holoprosencephaly) in Nueces County. Seven cases of anencephaly and two cases of holoprosencephaly were reported. Based on CBDMP rates, these were 7.1 times the expected number of anencephaly cases and 19.3 times the expected number of holoprosencephaly cases and statistically significant.

Response: TBDMD is in the process of confirming the reported cases and conducting an investigation to ascertain any additional cases for the county. The investigation is ongoing.

Duration of Investigation: May 1999 - December 1999

(14) OZONA (Crockett County)

Condition of Concern: *Cleft lip and cleft palate*

Time Period of Concern: August 1994 - March 1999

Background: A nurse notified TBDMD that she had seen three children with cleft lip and palate born in Ozona during 1994. Further information gathering revealed that two of the mothers actually resided in Ozona at the time of delivery. Since the town has a small number of births each year and both babies were born within a 4 month period, this was roughly 158 times what we would have expected based on rates from the CBDMP. This observation was statistically significant.

Response: To confirm the report and identify any additional cases, Texas Birth Defects Monitoring Division staff reviewed hospital medical records for all 44 deliveries to residents of Crockett County during 1994. The 2 cases initially reported among Ozona residents were confirmed, and no additional cases were detected among residents of the city of Ozona or Crockett County. Because there were not enough cases to make further investigation productive, the investigation was closed and a report was written. After receiving our report, the nurse who initially reported this cluster told us that 2 more children were born with cleft lip and palate during February and March 1999, and that the medical community continued to be concerned. The investigation was reopened and the time frame was extended to March 1999. The investigation is ongoing.

Duration of Investigation: September 1997 - December 1999

(15) PORT LAVACA (Calhoun County)

Conditions of Concern: *Gastroschisis*

Time Period of Concern: 1995 - 1998

Background: In February 1998, TBDMD was contacted by a nurse about a higher than expected number of gastroschisis cases in Port Lavaca. Three affected pregnancies were reported. Based on CBDMP rates, this was 14.2 times the expected number of cases and was statistically significant.

Response: TBDMD has confirmed the reported cases and is conducting an investigation to ascertain any additional cases for the county. The investigation is ongoing.

Duration of Investigation: February 1998 - December 1999

(16) PRESIDIO AND ALPINE (Presidio and Brewster Counties)

Condition of Concern: *Cleft lip and cleft palate*

Time Period of Concern: October 1997 - February 1998

Background: A concerned physician re-

ported 3 cases of cleft lip and palate in infants born from October 1997 through February 1998: 2 were residents of the city of Presidio, one was a resident of Alpine. The physician expressed concern over river water as a possible cause of these birth defects. Based on CBDMP rates, this number was roughly 55 times the expected total for the city of Presidio, 35 times the expected total for Presidio County, and 27 times the expected total for Presidio and Brewster Counties combined. These observations were statistically significant. Observed totals for Alpine and for Brewster County alone were not statistically significant.

Response: Since the geographic area and time period of concern are covered by the Texas Birth Defects Registry, registry data will be used to verify these reports and identify any additional cases among residents of Presidio County or Brewster County. Collection of registry data for 1998 is still underway, therefore the investigation is ongoing.

Duration of Investigation: April 1998 - December 1999

(17) ROBY (Fisher County)

Condition of Concern: *Multiple birth defects*

Time Period of Concern: Unknown

Background: A concerned parent contacted TBDMD regarding a concern about high rates of birth defects and other conditions in the area.

Response: Staff are working with the citizen to identify the geographic area, time period of interest, and the number of cases.

Duration of Investigation: December 1999

(18) SAN ANGELO AND BALLINGER (Tom Green and Runnels Counties)

Condition of Concern: *Hypoplastic left heart syndrome, tetralogy of Fallot, and transposition of the great arteries*

Time Period of Concern: June 1997 - December 1997

Background: A hospital nurse contacted TBDMD about four infants born with various heart defects and one infant with a collodian baby appearance. One child with a heart

defect also had an abdominal tumor. The heart defects reported included two cases of hypoplastic left heart syndrome, one case of tetralogy of Fallot, and one case of transposition of the great arteries.

Tetralogy of Fallot and transposition of the great arteries were categorized together as conotruncal heart defects. Based on rates from the CBDMP and the MACDP, the two cases of conotruncal heart defects were approximately 2 to 3 times what would be expected, but the findings were not statistically significant. The birth of 2 children with hypoplastic left heart syndrome was between 3 to 144 times what would be expected, and the findings were statistically significant for the city of Ballinger, Runnels County alone, and Tom Green and Runnels Counties combined.

Response: Since the geographic area and time period of concern are covered by the Texas Birth Defects Registry, registry data will be used to confirm reports and identify additional cases among residents of Tom Green or Runnels Counties. The investigation is ongoing.

Duration of Investigation: May 1998 - December 1999

(19) SMITH COUNTY

Conditions of Concern: *Down syndrome.*

Time Period of Concern: June 1998 - May 1999 (includes prenatally diagnosed Down syndrome)

Background: In September of 1998, TBDMD was contacted by a social worker about an apparent excess of cases of Down syndrome in Smith County. Nine affected pregnancies were reported. Based on CBDMP rates, this was 2.7 times the expected number of cases and statistically significant.

Response: TBDMD is conducting an investigation to (a) confirm reported cases and (b) ascertain and confirm any reported cases for the county. The investigation is ongoing.

Duration of Investigation: September 1998 - December 1999

(20) TOM GREEN COUNTY

Conditions of Concern: *Agenesis of the corpus callosum.*

Time Period of Concern: Unknown

Background: In October of 1999, TBDMD was contacted by a citizen of Tom Green County about what she had been told by her health care providers was a higher than expected number of agenesis of the corpus callosum cases in Tom Green County. Three cases were reported. Based on textbook reference rates, this was 12.2 times the expected number of cases and statistically significant.

Response: TBDMD is in the process of confirming the reported cases and conducting an investigation to ascertain any additional cases for the county. The investigation is ongoing.

Duration of Investigation: October 1999 - December 1999

(21) WEST DALLAS (Dallas County)

Conditions of Concern: *Birth defects (Eye defects)*

Time Period of Concern: Unknown

Background: In December of 1999, TBDMD was contacted by a member of the media about a higher than expected number of birth

defects (particularly eye defects) in West Dallas. A member of the community subsequently contacted TBDMD about the same cluster. Of particular concern was contamination from a lead smelting plant that

was closed down in the area in the 1980's.

Response: TBDMD is waiting for the community member to provide the number of cases and time period in an effort to perform initial calculations. The investigation is ongoing.

Duration of Investigation: December 1999

Acknowledgment: The Texas Birth Defects Monitoring Division is grateful for assistance during cluster investigations by other units of the Texas Department of Health, particularly the Texas Neural Tube Defect Surveillance and Intervention Project and the Environmental Epidemiology and Toxicology Division.

For more information about this report, contact the Texas Birth Defects Monitoring Division, 1100 W. 49th Street, Austin, TX, 78756. Phone 512-458-7232, Fax 512-458-7330. Additional information and previous years' cluster reports are available at www.tdh.state.tx.us/tbdmd/docs.htm.

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